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Sclerosing Angiomatoid Nodular Transformation (SANT) of the spleen: Case report and review of the literature

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ABSTRACT

INTRODUCTION: Sclerosing Angiomatoid Nodular Transformation of the spleen (SANT) is a rare benign vascular lesion of the spleen with extensive sclerosis and unknown etiology.
PRESENTATION OF CASE: We report a new case of SANT of the spleen found in a 53-year-old female following detection of a splenic mass on a routine computed tomography (CT). The patient underwent an uncomplicated laparoscopic splenectomy and the specimen was sent for histopathologic examination.
DISCUSSION: A review of the 97 reported cases of SANT found in the literature was undertaken. There were 43 males and 54 females with a median age of 46 years (range: 11–82 years). SANT is classically considered to be a female predominant disease, however 44.3% of reported case were male and the gender predilection may soon be neutralized as more cases are reported. 65 of the 97 (67%) patients were in 30–60 year age group. The majority of lesions ($n = 50$) were incidentally found on imaging, and for those patients presenting with symptoms, abdominal pain ($n = 18$) was the predominant symptom.
CONCLUSION: The diagnosis of SANT should be considered in any patient presenting with a splenic lesion that contains an angiomatoid or inflammatory component. As the differential diagnosis for SANT includes malignant pathologies, and currently no reliable diagnostic radiological feature has been identified to differentiate between these conditions, SANT will continue to be diagnosed on the basis of surgical histopathology.

1. Introduction

Sclerosing Angiomatoid Nodular Transformation (SANT) of the spleen is a rare benign vascular lesion with extensive sclerosis first described by Martel and colleagues in 2004.[1] In this paper we report a case of SANT of the spleen managed at our institution, and present a review of 97 other cases found in the literature.

2. Presentation of case

A 53 year old Caucasian female was referred to our department in consideration of splenectomy. She had been under the care of the hematology department following the detection of a splenic mass on a routine computed tomography (CT) scan performed for chronic back pain (Fig. 1). An ultrasound scan at that time demonstrated a $3.6 \times 3.5 \times 3.5$ cm hypoechoic splenic density, and a magnetic resonance imaging (MRI) scan (Fig. 2) confirmed the mass within the inferior spleen demonstrating diffuse heterogeneous enhancement. At this time the differential diagnosis included Gaucher's disease, sarcoid or a low-grade lymphoma. Bone marrow biopsy, flow cytometry, chromosome analysis and angiotension converting enzyme levels were normal, and the above differential diagnoses ruled out.

Over a three year period the splenic mass increased in diameter to 5.9 cm and the referring hematologist was concerned for the possibility of an underlying malignancy. The patient remained asymptomatic and her examination benign, with no evidence of hepatosplenomegaly. The decision was made to proceed with an operation and the patient underwent a laparoscopic splenectomy.

2.1. Intraoperative findings

On entering the abdominal cavity significant adhesions were encountered in the region of the inferiolateral aspect of the spleen where the mass was clearly visible. When the spleen was placed into a bag for morcellation prior to extraction, it was noted that the mass was significantly harder than the rest of the spleen. As a result the surrounding normal splenic tissue was morcellated until only the mass was remaining, and this was then removed in its entirety and sent for histopathological examination.

2.2. Pathologic findings

The splenic mass was composed of mutiple nodules of small vessels surrounded by sclerotic tissue and a scattered lymphoplasmacytic infiltrate. Immunohistochemical staining of the small

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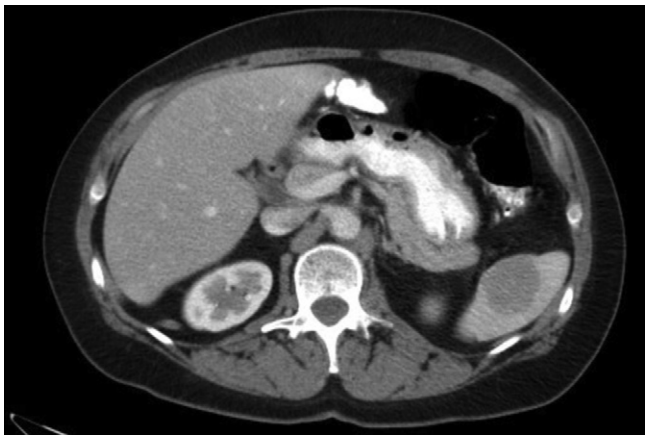

Fig. 1. Computed tomography (CT) scan showing the splenic lesion at the medial aspect of the lower spleen.

vessels within the lesion were positive for CD34, CD31, CD68 and CD163, and negative for CD8 (Figs. 3–7). The histologic sections and immunohistochemical staining performed on the splenic lesion confirmed the diagnosis of SANT of the spleen.

3. Discussion

The term SANT first appeared in the literature in a 2004 paper by Martel et al. which examined a series of 25 cases.[1] This relatively uncommon splenic lesion had however been recognized earlier by other authors under different names such as splenic hamartoma, cord capillary hemangioma, and multinodular hemangioma.[2] SANTs are benign, nodular vascular proliferations of splenic red pulp with considerable sclerosis.[1,26] It usually affects middle-aged adults, and it is commonly found incidentally on radiographic imaging, or at the time of operation for an unrelated condition.[20]

Review of the existing literature revealed 97 patients consisting of 43 males and 54 females with a median age of 46 years (range: 11–82 years). SANT is considered to be a female predominant disease,[26] however 43 of the 97 cases (44.3%) reported in the literature to date were male and the gender predilection may soon be neutralized as more cases are reported. 65 out of the 97 (67%) patients were in 30–60 year age group, so it appears that SANT predominantly affects adults in the fourth to seventh decades of life. The majority of lesions ($n = 50$) were incidentally found on

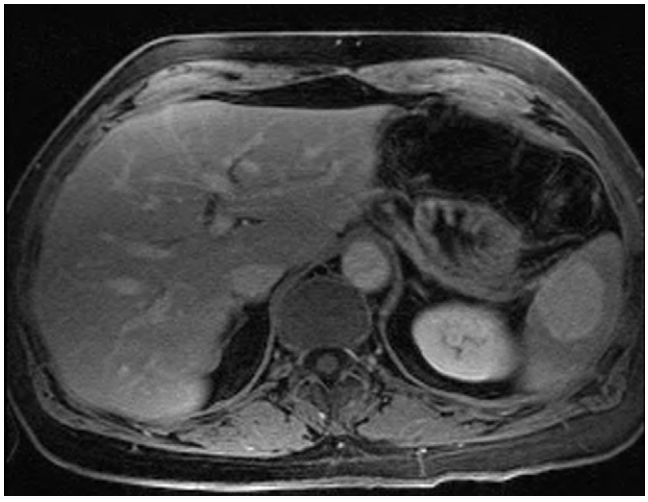

Fig. 2. Magnetic resonance imaging (MRI) showing the lesion in the lower spleen.

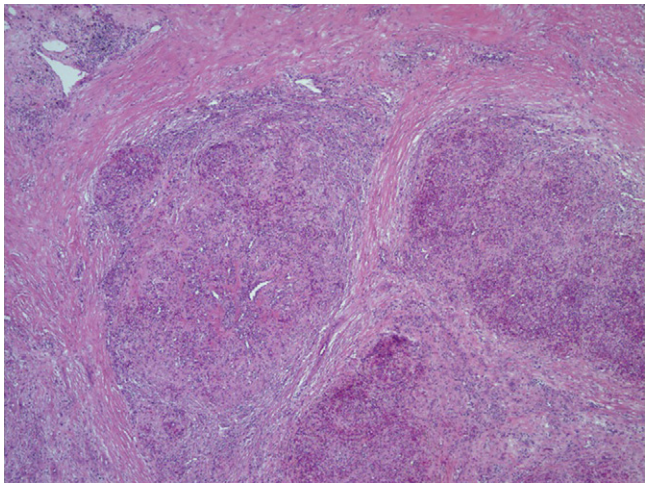

Fig. 3. The splenic parenchyma is replaced by innumerable well-circumscribed angiomatoid nodules separated by a fibrosclerotic and inflammatory stroma. The nodules are composed of a variety of cell types including capillaries, sinusoid-like spaces, and mononuclear inflammatory cells. Red blood cells are abundant.

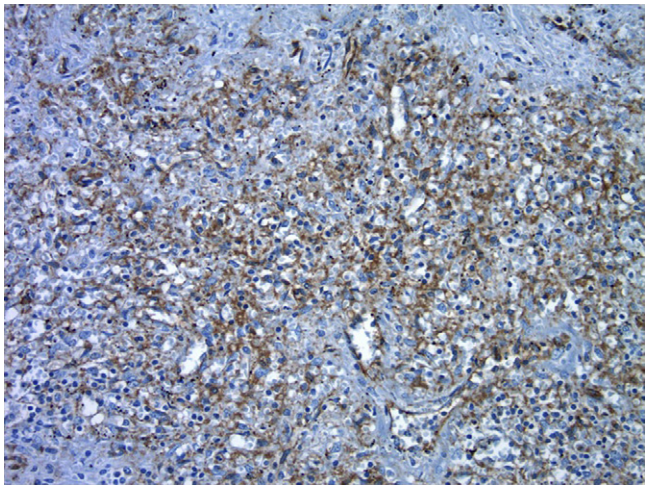

Fig. 4. CD31 immunostain highlights the abundant vascular structures (capillaries, sinusoid-like spaces, and veins) along with numerous single cells within the nodules, generating a complex network of CD31 immunoreactive cells.

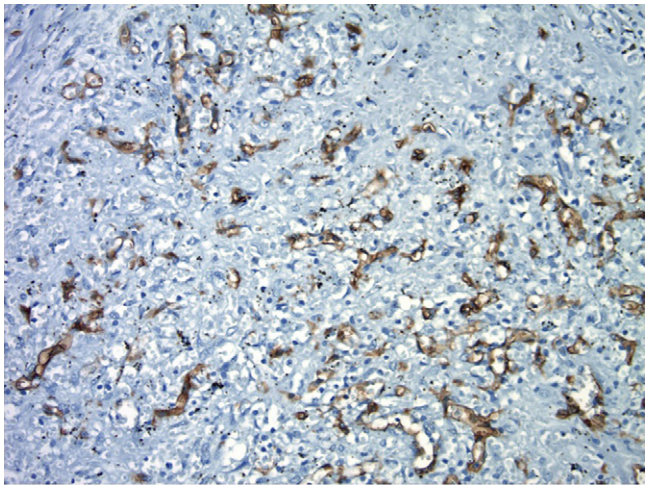

Fig. 5. CD34 immunostain highlights the capillaries, but not sinusoid-like spaces or any single cells.

Table 1
Published SANT cases.

	Author	Age	Gender	Clinical features	Spleen weight	Gross features	Follow-up	Referring dx	Concurrent disease
1	Martel[1] *N* = 25	50	Female	Incidental finding at laparotomy	329 g	–	NED, 9 years	–	
2		32	Female	Incidental radiographic finding	139 g	5 cm mass with white fibrous bands	NED, 8 years	Hamartoma	
3		57	Male	Incidental radiographic finding	–	3.7 cm fibrotic mass	NED, 6 years	Hemangioma vs. angiosarcoma	
4		58	Female	Incidental radiographic finding	456 g	9 cm firm mass	NED, 5 years	Hemangioma vs. IPT	
5		35	Female	Pancytopenia, raised ESR	–	8 cm well-circumscribed mass	NED, 4 years	Hemangioma	
6		71	Female	Incidental radiographic finding	704 g	13.5 cm well-circumscribed fibrotic mass	NED, 4 years	Hemangioma vs. IPT	
7		23	Male		280 g	7 cm well-circumscribed fibrotic mass	NED, 3 years	–	
8		59	Female	Incidental finding at laparotomy	130 g	4 cm fibrotic mass	NED, 4 years	Angiosarcoma vs. IPT	
9		37	Female	Abdominal pain	280 g	6 cm mass	NED, 1 year	Hemangioma vs. littoral cell angioma	
10		29	Male	Abdominal pain	–	10 cm mass	NED, 2 years	Hemangioma vs. IPT	
11		60	Female	–	1400 g	12 cm mass	NED, 10 m	–	
12		74	Male	Incidental finding at laparotomy	160 g	3 cm well-circumscribed multilobulated mass	NED, 6 m	Hemangioma	Renal cell carcinoma
13		57	Female	Incidental finding at laparotomy	173 g	8 cm mass	NED, 6 m	Vascular tumor	
14		61	Female	Splenomegaly	68 g	3.5 × 3.5 cm sharply demarcated, gray-white to red-purple mass	–	–	
15		56	Female	Abdominal discomfort, anemia	189 g	5.5 cm well-circumscribed mass with red-brown nodules traversed by stellate fibrous bands	NED. Died of lung ca	IPT	
16		46	Male	Hx of anemia and lung SCC. Presented with fever and splenomegaly	–	–	Died of sepsis post splenectomy	Bacillary angiomatosis vs. Kaposi sarcoma	
17		68	Female	Carcinoma of colon 8 m prior with post-op chemo. Incidental finding	285 g	4.5 cm bosselated whitish mass surrounded by brown nodules	NED, 7 years	Benign Kaposi-like vascular tumor vs. bacillary angiomatosis vs. chemo-related changes	
18		63	Male	RUQ pain. Incidental finding	93 g	4 cm mass	NED, 3 years	–	Early gastric carcinoma
19		56	Male	Incidental finding	–	10 cm circumscribed whitish mass with focal hemorrhage	–	IPT	
20		25	Female	LUQ mass	335 g	8.5 × 6 × 5 cm circumscribed, grayish mass	NED, 3 years	IPT vs. sclerosedhemangioma vs. Castleman disease	

Table 1 (*Continued*)

	Author	Age	Gender	Clinical features	Spleen weight	Gross features	Follow-up	Referring dx	Concurrent disease
21		45	Male	Incidental finding	322 g	circumscribed firm, gray-white nodule with brown patches	NED, 3 yearss	Hemangioendothelioma vs. IPT	
22		43	Female	Incidental finding	250 g	–	NED, 2 m	N/A	
23		35	Female	LUQ pain for 6 months	240 g	3.1 × 2.5 cm subcapsular mass, pale, sclerotic in center and brown-red at periphery	–	Hemangioepithelioma vs. Kaposi sarcoma	
24		23	Female	Palpable mass	1425 g	17 × 11 cm irregular fibrous mass with thick capsule	NED, 1 year	Sclerosedhemangioma vs. hamartoma	von Willebrand disease
25		57	Female		105 g	3.5 × 3.4 × 3.2 cm mass with multiple dark red nodules	NED, 18 m	Hemangioma	
26	Li[3] *N* = 1	59	Male	Incidental finding on CT during workup for renal problems	283 g	3.3 × 3.3 cm firm yellow mass			HTN, DM, hypothyroidism, BPH
27	J.-C. Lee[10] *N* = 1	43	Female	Weight loss and left flank pain	180 g	3.5 × 3.5 × 3.0 cm and 3.0 × 2.0 × 2.0 cm Hemorrhagic nodules		Thrombosedhemangioma, lymphoma, chronic abscess	Hepatitis B+
28	D. Lee[4] *N* = 1	58	Male	Incidental finding on U/S. Deranged LFTs	205 g	8.7 × 6.5 × 5.5 cm		Metastatic melanoma to spleen	Previous malignant melanoma
29	Weinreb[5] *N* = 6	58	Female	Lung Ca staging CT	110 g	1.9 cm	NED, 1 year	SANT	
30		65	Male		320 g	6.5 cm	Lost to f/u	Hamartoma	
31		73	Female	Hypertension, hyperlipidemia, remote lung abscess, chronic cough	324 g	6.0 cm	NED, 16 m	Hemangioendothelioma	
32		51	Male	Anemia	2720 g	12.0 cm	NED, 5 m	IPT	
33		41	Female	Enlarging mass on serial U/S	256 g	6.5 cm	NED, 1 m	SANT	
34		59	Female	Multiple splenic lesions 20 years post Whipple for periampullary Ca	–	2.1 cm	NED, 1 year	IPT	
35	Diebold[11] *N* = 16	56	Female		2400 g	3 nodules 1.0, 2.0 and 3.0 cm			Idiopathic myelofibrosis
36		22	Female		220 g	5.5 × 3.0 cm multinodular			Acute pyelonephritis
37		37	Female		590 g	10.0 × 7.0 cm multinodular			
38		33	Female	Hypochromic anemia	1760 g	multinodular			
39		60	Female		430 g	10.0 cm multinodular			
40		44	Male	Longstanding fever	610 g	15.0 cm multinodular			
41		24	Female	Hypochondral pain	160 g	5.0 cm multinodular			
42			Male	Gastric ulcer	218 g	1.0 cm			
43		31	Female		180 g	7.0 cm multinodular			
44		46	Male	Thrombocytopenia	137 g	4.0 cm multinodular			
45		82	Male		–	2.0 cm multinodular			Colon carcinoma with mets to splenic hilum

Table 1 (*Continued*)

	Author	Age	Gender	Clinical features	Spleen weight	Gross features	Follow-up	Referring dx	Concurrent disease
46		53	Male		190 g	5.0 cm multinodular			
47		63	Female	Fever, night sweats	140 g	1.5 × 1.0 × 1.0 cm multinodular			
48		55	Male	Anemia	550 g	8.0 cm multinodular			
49		62	Female	Abdominal pain		4.0 cm multinodular			
50		50	Male	Anemia	840 g	9.0 cm multinodular			
51	El Demellawy[12] *N* = 1	58	Female	Incidental finding. Hx of RUL non-small cell carcinoma stage III	110 g	1.9 cm well-circumscribed but non-encapsulated nodule. Bosselated contour and whitish, firm, solid surface		Metastatic lung cancer	
52	Karaosmanoglu[13] *N* = 1	44	Male	Vague pelvic pain	650 g	Whitish firm nodule with hemorrhagic spiculations			
53	Zeeb[14] *N* = 1	36	Female	LUQ Pain for 2 weeks		Multiple red-brown nodules with a prominent stellate scar			
54	Teng[15] *N* = 1	37	Female	Incidental finding with hepatolitiasis	1080 g	8.5 × 8.5 cm			
55		31	Male	Incidental finding	780 g	8.0 × 8.0 cm			
56		58	Female	Flank/back pain for 1 month	528 g	6.5 × 4.5 cm			
57		31	Female	Upper abdominal discomfort × 2 years	2106 g	8.5 × 8.5 cm		SANT	
58		37	Male	Incidental finding	297 g	3.0 × 3.0 cm		SANT	
59		37	Male	Incidental finding	473 g	3.5 × 3.0 cm		SANT	
60		50	Female	Incidental finding with hepatic angioma	314 g	2.6 × 2.4 cm		SANT	
61	Kashiwagi[16] *N* = 9	31	Female	Incidental Finding	230 g	5.5 cm	–	IPT	
62		34	Male	Back discomfort	–	7.0 cm	NED, 3 m	IPT	
63		37	Male	Epigastric pain	80 g	3.0 cm	NED, 79 m	IPT	
64		44	Female	Incidental Finding	–	3.5 cm	NED, 25 m	SANT	Cholelithiasis
65		46	Male	Incidental Finding	110 g	6.5 cm	–	IPT	Chronic hepatitis
66		50	Male	Incidental Finding	500 g	11.0 cm	–	IPT	
67		60	Male	Incidental Finding	100 g	2.5 cm	NED, 37 m	SANT	Gastric cancer
68		65	Female	Incidental Finding	–	5.1 cm	–	IPT	Cholelithiasis
69		72	Female	Incidental Finding	–	2.0 cm	NED, 113 m	IPT	Colon cancer
70	Gutzeit[17] *N* = 1	77	Male	Incidental finding on CT. Patient had prostate cancer		8 × 6 cm		Hamartoma	
71	Koreishi[6] *N* = 3	58	Female	Abdominal pain	307 g	4.4 cm in inferior pole	NED; Alive		DM, hypothyroidism

Table 1 (*Continued*)

	Author	Age	Gender	Clinical features	Spleen weight	Gross features	Follow-up	Referring dx	Concurrent disease
72		72	Female	Incidental finding or annual CT scan	79 g	2.3 cm	NED; Alive		Hx of high-grade urothelial carcinoma of renal pelvis and low-grade urothelial carcinoma of bladder
73		64	Female	Incidental finding on routine CT scan	110 g	2.1 cm	NED; Alive		Hx of carcinoma of fallopian tube, malignant melanoma in situ
74	Langer[18] *N* = 1	44	Male	Incidental finding on routine CT scan	170 g	2.0 cm mass; encapsulated reddish nodular	NED 4 m; Alive	Metastatic rectal cancer	
75	Chikkappa[19] *N* = 1	40	Female	Intermittent LUQ pain	168 g	4.7 × 4.0 × 6.5 cm peripheral circumscribed nodule, which was partly fibrous and partly nodular hemorrhagic		SANT	
76	Thacker[20] *N* = 1	80	Male	Incidental radiographic finding	–	Granular and gray-purple with a lobulated 9 cm mass with hemorrhagic areas measuring 0.1–0.5 cm		SANT	MDS, Melanoma, Basal cell carcinoma and squamous cell carcinoma
77	Kuybulu[21] *N* = 1	11	Female	Incidental finding on physical examination		Well circumscribed confluent vascu-lar/angiomatoic nodules with mixed-type inflammatory cells	NED, 1 year	SANT	Short stature
78	Kuo[22] *N* = 10	32	Female	Left flank soreness for 2 weeks	139 g	Single 3 × 4 × 5 cm nodule	NED, 94 m	SANT	
79		53	Female	Diffuse abdominal pain	–	Single 3 × 3 cm nodule	NED, 166 m	SANT	
80		57	Female	Incidental finding	105 g	Single 3.5 × 3.4 × 3.2 cm nodule	–	SANT	
81		37	Male	Incidental finding	275 g	Single 6 × 6 cm nodule	–	SANT	
82		46	Female	RUQ pain for 2 months	104 g	Single 2.2 × 2 × 2 cm nodule	–	SANT	
83		39	Male	Incidental finding	278 g	Multinodular	NED, 14 m	SANT	
84		31	Male	Right inguinal mass and Incidental radiographic finding	654 g	Multinodular	NED, 6 m	SANT	
85		57	Male	Left upper abdominal pain for 2 years	142.5 g	Multinodular	–	SANT	
86		33	Female	LUQ Pain for 1 year	143.6 g	Single 5.2 × 5 × 4 cm	NED, 2 m	SANT	
87		44	Male	Incidental finding	212.6 g	Single 6.8 × 6 × 4.5 cm	NED, 1 m	SANT	

Table 1 (*Continued*)

	Author	Age	Gender	Clinical features	Spleen weight	Gross features	Follow-up	Referring dx	Concurrent disease
88	Cao[23] *N* = 3	36	Male	Incidental radiographic finding	–	Mass was found to have an integrated envelope and a heterogenous cut surface		SANT	
89		37	Female	Pain in the LUQ	–	Firm mass with a clear margin		SANT	
90		39	Male	LUQ mass	–	–		SANT	
91	Sitaraman[24] *N* = 1	65	Male	Incidental radiographic finding	750 g	2 cm well-circumscribed nodule with an area of central fibrosis		SANT	Retroperitoneal spindle cell sarcoma
92	Subhawong[25] *N* = 1	27	Female	RUQ pain after motor vehicle crash	474 g	10.2 cm firm, white, nodular lesion infiltrating red irregularly		SANT	Unexplained anemia with history of transfusion
93	Bamboat[26] *N* = 1	17	Male	Abdominal pain for 6 months		Lobulated 4 cm fibrotic mass with hemorrhagic areas	NED, 7 m	SANT	
94	Raman[27] *N* = 1	50	Male	LUQ pain for 4 months					
95	Ki-Han[28] *N* = 1	23	Female	Incidental radiographic finding	–	5.2 × 4.5 cm dark brown mass with a central large stellate fibrotic scar		SANT	
96	Onder[29] *N* = 1	48	Male	Pelvic pain	650 g	8 cm solitary non-encapsulated mass composed of multiple nodules with wide area of hemorrhage and a central stellate scar		SANT	
97	Vyas[30] *N* = 1	11	Male	Left flank pain since 2 months	125 g	5 × 4 × 4 cm well-circumscribed unencapsulated lesion with bulging cut surface and central fibrotic scar	NED, 3 years	SANT	

imaging, and for those patients presenting with symptoms, abdominal pain ($n = 18$) was the predominant symptom. Other presentations included: a palpable left upper quadrant mass; cytopenias; flank pain; pelvic pain; and long-standing fever.

The weight of resected spleens in the literature exhibited significant variation from 68 to 2720 g. The typical macroscopic appearance of a SANT lesion was of a well-circumscribed non-encapsualted, bosselated mass with multiple dark brown nodules (hemorrhagic regions in angiomatoid nodules) interspersed with stellate whitish fibrotic stroma.[1,26,27] The cases reported before 2008 had varying diagnoses that included hamartoma, inflammatory pseudotumor, hemangioma, angisarcoma, metastatic tumor, bacillary angiomatosis, but thereafter SANT has been the referring diagnosis.

There is minimal data available on the follow-up of patients with SANT. There are two reported deaths in the 25 cases published by Martel et al.[1]; a 56-year-old female who died of disseminated lung adenocarcinoma, and the other a 46-year-old male with concurrent bronchogenic squamous cell carcinoma who died of sepsis post-splenectomy. There is no data regarding the immunization status and use of antibiotic prophylaxis in these patients (Table 1).

There is currently no pathognomonic finding for the diagnosis of SANT on cross-sectional imaging, however, the literature suggests that the diagnosis can be made if a contrast-enhanced MRI shows a "spoke-wheel pattern".[20,26] Gutzeit et al.[17,23] propose the use of contrast-enhanced ultrasonography (CEUS) to diagnose SANT, but the role of CEUS needs to be further evaluated as data is limited. There have been two reports of F-18 fluorodeoxyglucose (FDG)-avid splenic lesions found to be SANT lesions,[20] however other authors have reported SANT cases without PET activity.[6]

Martel et al. found three distinct types of blood vessels in the specimens they examined, mirroring the normal composition of splenic red pulp.[1] The first were well-formed cord capillaries in an organized lobular arrangement that were CD34+/CD8−/CD31+. The second type of vessel were consistent with splenic sinusoids and were CD34−/CD8+/CD31+. The third type consisted of small

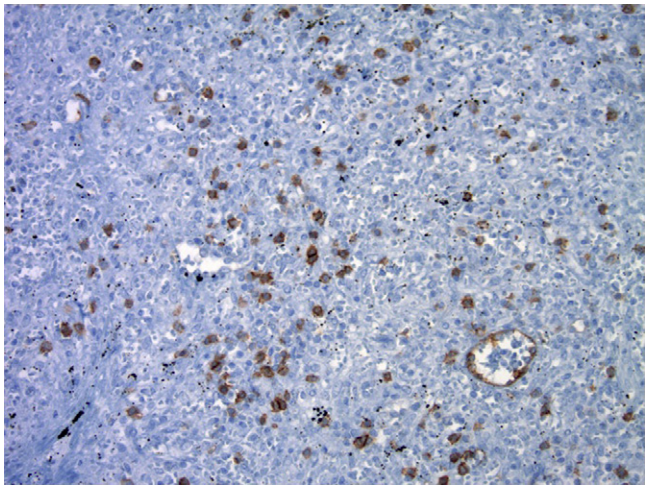

Fig. 6. CD8 immunostain highlights occasional sinusoid-like spaces (lower right) and scattered inflammatory cells, but is absent in other vascular structures (capillaries and veins).

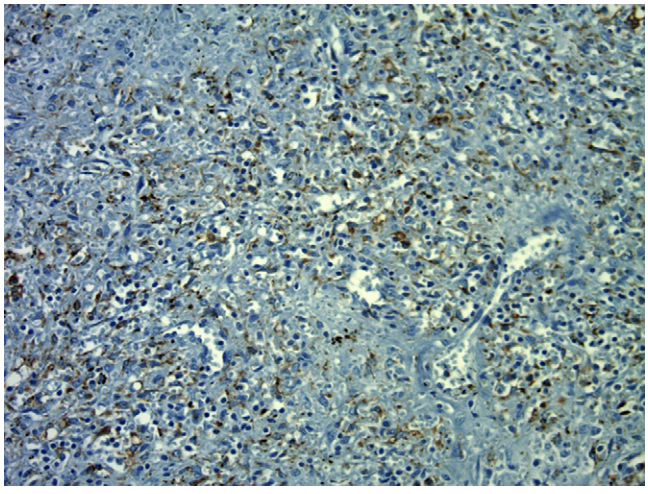

Fig. 7. CD68 immunostain highlights scattered single cells (presumably histiocytes) but no vessel-lining cells.

veins arranged in a very intricate mesh-like patterns, and were CD34−/CD8−/CD31+. The nodules of vessels are separated by collagenous bands, and the stroma between nodules is sclerotic.

As SANT is a vascular lesion comprised of an over-proliferation of blood vessels, its differential diagnosis includes other benign lesions such as hamartomas, hemangiomas, hemangioendotheliomas, littoral cell angiomas, or inflammatory myofibroblastic lesions. Martel et al. noted that the pathogenesis of this entity is unclear and hypothesize that SANT may be a splenic hamartoma that has undegone an unusual form of sclerosis, with a peculiar reactionary transformation of red pulp due to an exaggerated stromal response.[1] It appears that SANT is probably a reactive lesion rather than a true neoplastic process, a theory supported by the high prevalence of concurrent conditions in SANT patients.

The fact that SANT can resemble an inflammatory pseudotumour has prompted some authors to suggest that the two lesions may in fact be the same.[5] In support of this hypothesis there have been reports of SANT cases which show EBER-1 (Epstein–Barr virus-encoded small RNAs) positive stromal cells.[5] However, while the stroma of IPT and SANT may be histologically similar, IPT do not contain the angiomatoid nodules seen in SANT.[8] Recently a number of authors have suggested that the proliferation seen in SANT may be related to IgG4 sclerosing lesions due to the presence of plasma cells found in its stroma.[6,7]

As this lesion is benign without risk of malignant transformation, the question arises whether an asymptomatic patient with SANT should undergo an operative procedure if the lesion is found incidentally? There is currently no sensitive and specific way to make a diagnosis of SANT without having a tissue sample, and as some lesions that resemble SANT are malignant in nature, we think it prudent to operate even if SANT is suspected. Core biopsy is a sensitive and specific way to diagnose both hematologic and non-hematologic splenic lesions.[8,9] Weinreb et al. argue that due to its distinctive nodular pattern, lack of atypia, and unique immunohistochemical profile, that core biopsy can be used to distinguish SANT from other lesions in the differential diagnosis of SANT.[5] However, an important factor which Weinreb et al. do not appear to consider, is the risk of intra-peritoneal seeding if the lesion being biopsied proves to be say an angiosarcoma.[5]

4. Conclusion

The diagnosis of SANT should be considered in any patient presenting with a splenic lesion that contains an angiomatoid or inflammatory component. There is a wide age distribution and the gender distribution appears to be equal. The majority of cases of SANT reported in the literature were incidental diagnoses, with the remainder presenting with a variety of non-specific symptoms. As the differential diagnosis for SANT includes malignant pathologies, and currently no reliable diagnostic radiological feature has been identified to differentiate between these conditions, SANT will continue to be diagnosed on the basis of surgical histopathology.

Conflict of interest statement

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Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contributions

Gavin A. Falk – manuscript design, data collection, writing; Nishank P. Nooli – data collection, writing; Gareth Morris-Stiff – data collection, writing; Thomas P. Plesec – pathology review and writing; Steven Rosenblatt – manuscript design, writing.

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